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Vitiligo colocalising with Kyrle's disease: A peculiar manifestation

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Abstract

A rare form of acquired perforating dermatosis (APD) called Kyrle's disease (KD) is linked to several systemic illnesses, including chronic kidney disease and diabetes mellitus (DM). The lower extremities are the sites where it most frequently occurs.

We report here a case of 49-year-old diabetic female who presented to our Skin OPD with multiple hyperkeratotic papules & nodules over B/L lower limbs with depigmentation at the base. A diagnosis of Kyrle's disease colocalised on vitiligo was made. The diagnosis of Kyrle's disease was confirmed after Skin biopsy. Vitiligo is a common autoimmune disease targeting the skin, with Th1 cells implicated in the pathogenesis. High potency topical steroids added with keratolytic, and antibiotic doxycycline was prescribed which showed good response.

Keywords: APD, Kyrle's disease (KD), diabetes mellitus (DM).

Introduction

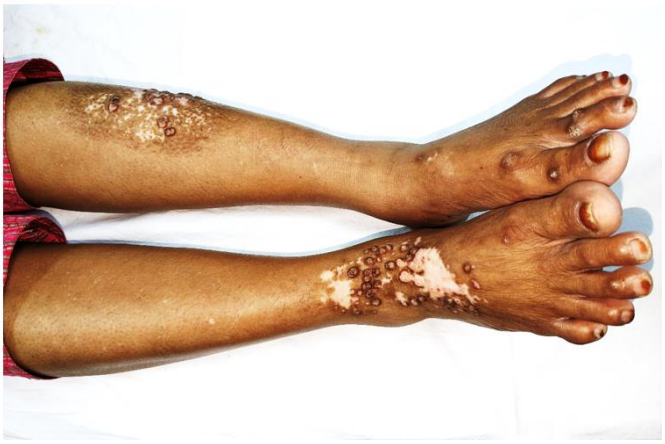
A rare skin condition known as Kyrle's disease (KD) is characterized by the trans epidermal removal of aberrant keratin. Patients with diabetes, kidney disease, and liver illness are the most likely to experience it.[1] KD is thought to be a genetically determined disease that often manifests in maturity between the ages of 30 and 50, but it has also been documented to manifest as early as age 5 and as late as age 75 [2], with a female-to-male ratio of up to 6:1.[3]Often on the lower extremities, it manifests as a number of distinct, eruptive papules with a central crust or plug. Keratolytics (salicylic acid and urea), electrocautery, or radio-cautery are the first-line treatments for KD. A prevalent depigmenting skin condition called vitiligo is thought to affect 0.5–2% of people worldwide. Melanocytes are selectively lost in

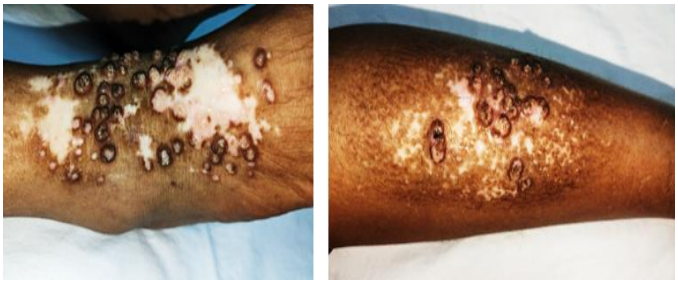
the condition, resulting in the distinctive non-scaly, chalky-white macules.[4] Colocalization of both conditions warrants' investigation.

Case Report

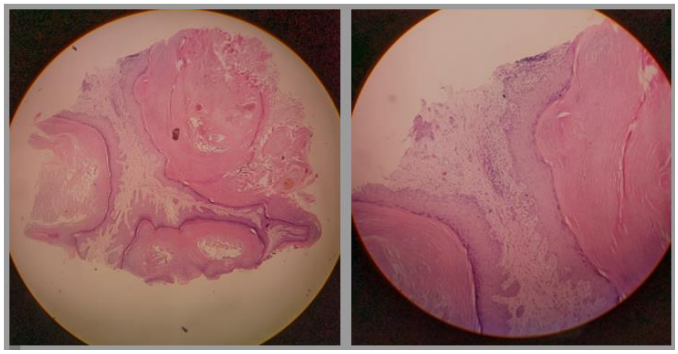
49-year female patient presented in Dermatology Opd with chief complaints of multiple elevated lesions over bilateral lower limbs for 1.5 years which were associated with itching. Depigmentation was seen at the base of all these elevated lesions. Patient took treatment for these lesions from local general practitioners but didn't get relief. The patient was a known case of Type 2 Diabetes Mellitus and has been taking antidiabetics for 15 years. A general physician was consulted for management of the same.

On local examination, multiple hyperkeratotic papules and nodules were present over anterior aspect of B/L lower limbs which were itchy in nature disturbing patient's day to day activities. A detailed medical history, physical examination and blood investigations did not reveal any comorbidities. So, Skin biopsy was advised for confirmation.





Clinical pictures showing multiple hyperkeratotic papules present over anterior aspect of bilateral lower limbs with a central keratotic plug.



Histopathology showing shallow extrafollicular cup-shaped epidermal invagination filled with compact orthokeratosis and dermis showing dense inflammatory infiltrate.

Patient was treated with topical steroids, emollients, keratolytic agents and antibiotic doxycycline. Anti histamines were prescribed to alleviate the itching. There is slow but noticeable improvement in the lesions. Patient was counselled about the chronic, relapsing nature of the disease and therefore, need for regular follow up.

Discussion

The exact etiology of Kyrle's disease has not been elucidated. According to widely accepted case reports, it usually develops because of chronic systemic disorders, most frequently diabetes mellitus and stage 4 or stage 5 renal failure. When there is a female-to-male ratio of up to 6:1, the incidence is higher in females. Kyrle's disease is diagnosed based on characteristic clinical and histopathologic findings. It typically presents as

hyperkeratotic papules and nodules with a central keratotic plug.

The lower extremities, particularly the calf, tibial region, and posterior aspect, are where skin lesions are most frequently found. The head, neck, and arms may also be affected. There have also been reports of Koebnerization, in which skin lesions develop at the areas of damage. According to histology, the hallmark of Kyrle's disease is transdermal removal of keratotic material devoid of collagen or elastic fibers. It stands out from other acquired perforating illnesses since it doesn't eliminate collagen and elastic fibers via the skin. Additionally, the epidermis has an atrophic appearance and an underlying keratotic plug. There could be a dermal histiocytic and lymphocytic infiltration as part of a foreign body granulomatous reaction. Additionally, aberrant keratinization, orthokeratosis, and parakeratosis may be seen. Currently, treatment suggestions are based on case studies or anecdotal information.

Salicylic acid and urea are examples of keratolytics that are used as first-line treatments. For the treatment of pruritus, emollients and oral antihistamines have been employed. Alternative treatments include electrocautery, cryotherapy, CO2 laser, topical retinoids, isotretinoin, and psoralen combined with ultraviolet A radiation. Surgical excision is only used in severe, refractory cases where the skin lesions have not responded to other treatments.

Primary, restricted, or generalized cutaneous and mucosal depigmentation known as vitiligo is influenced by genetics, melanocyte self-destruction, cytokines, autoimmune, and oxidative stress. [5]

While more research is still needed to understand the precise chemical pathways. Recent research has demonstrated that the IFN-gCXCL9/10-CXCR3 axis may play a role in the development of vitiligo by suppressing melanogenesis, causing melanocyte apoptosis, and further attracting T lymphocytes to the skin. All of these contribute to the JAK/STAT pathway.[6]

KD has been seen in association with multiple disorders, including diabetes mellitus, renal and liver diseases, congestive heart failure, hyper-lipidaemia, infective diseases, and abnormal metabolism of vitamin A. Association with vitiligo is scarce in literature and needs further research.

Conclusion

When determining the differential diagnosis of erythematous papules with a central depression and keratotic plug, Kyrle's illness should be considered. KD is a rare condition that has the potential to lower quality of life. Therefore, when treating patients with diabetes and concurrent renal illness, doctors need to have a high degree of suspicion for KD. Any suspicious lesion should be biopsied, sent for histology, and treated as soon as possible after the diagnosis has been made. Since there is little literature on KD connections, more extensive research is required to better understand the condition and, ultimately, improve patient outcomes.

Declaration of Patient

In their statement of patient consent, the authors state that they have the necessary patient permission documents on file. The patient has indicated in the form that he is fine with images and other clinical data being published in the publication. He understands that, while every attempt will be made to conceal his identity and that his name and initials will not be published, anonymity cannot be guaranteed.

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